# CASE REPORTS

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# Vaginal Ectopic Ureter: A Continuing Diagnostic Challenge

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ECTOPIC URETER DRAINING to the vagina is a rare congenital malformation that causes significant symptoms and morbidity and is rarely considered early. The symptomatology is pathognomonic but variable. In the cases reported here, vaginal discharge and chronic perineal wetness were the major symptoms, rather than the more classic urinary incontinence. In any case where this entity is clinically suspected, aggressive workup should begin with intravenous urography, followed by ultrasonography and vaginography. Depending on results of ultrasonogram or vaginogram further workup should be pursued.

## Reports of Cases

CASE 1. A 4-year-old girl presented with a history of urinary frequency, "no bladder control" and recurrent yellowish vaginal discharge since toilet training, which had occurred at age 3. Voiding cystourethrography (VCU) elicited no abnormality. Intravenous urography (IVU) showed a normal right kidney but nonvisualization on the left. At cystoscopy a left ureteral orifice could not be identified. Over the next two years the patient had multiple febrile illnesses that were often associated with left abdominal pain. Voiding cystourethrography, intravenous urography and cystoscopy were repeated with no change in the findings. At 6 years of age, intravenous urography with tomography failed to demonstrate a left kidney.

Because of suspicion of an ectopic ureter draining a small left kidney, an aortogram was done, which failed to show a left renal artery. Through the ensuing four years the patient continued to have perineal wetness, recurrent vaginal discharge and recurrent episodes of fever and left flank and left abdominal pain. At 10 years of age a vaginogram (Figure 1) showed retrograde filling of a left ureter and renal collecting structures.

The patient subsequently underwent a left nephroureterectomy. At operation the left kidney was very small (3 by 2 by 1 cm) and there was duplication of the collecting structures. The lower pole portion of the kidney was very hypoplastic and its ureter was atretic in its proximal portion. The upper pole collecting system drained ectopically to the apex of the vagina. Histologic examination revealed renal dysplasia and chronic inflammatory change. During the ensuing six years the patient has done well with no further perineal wetness or vaginal discharge.

CASE 2. A 7-year-old girl presented with a four-year history of vaginal discharge and persisting perineal wetness. These complaints had been evaluated by at least five different physicians, one of whom had treated her for gonorrhea. The patient had had episodes of left lower abdominal pain but none were associated with fever. Intravenous urography (Figure 2) revealed a so-called cystic area in the upper pole of the left kidney; the collecting structures of the lower pole area appeared normal but displaced by the upper pole cystic mass. An ectopically draining ureter was suspected, so a vaginogram was done but failed to show the suspected findings. Ultrasonography demonstrated a sonolucent area in the left upper pole. A 20-gauge sheathed needle was inserted percutaneously in this cystic area and antegrade pyelography was done (Figure 3), demonstrating a duplicated collecting system with the upper pole portion draining ectopically into the vagina. Methylene blue instilled into this system appeared in the vagina in 30 minutes. Subsequent cystoscopy was normal and vaginography under anesthesia did not show the ectopic ureteral orifice. A left

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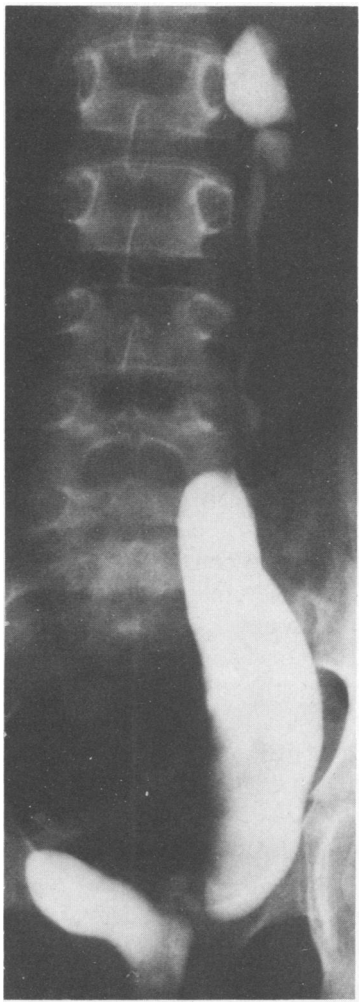

**Figure 1.**—Vaginogram from Case 1: The dilated distal left ureter and dilated duplicated collecting system are opacified with contrast material.

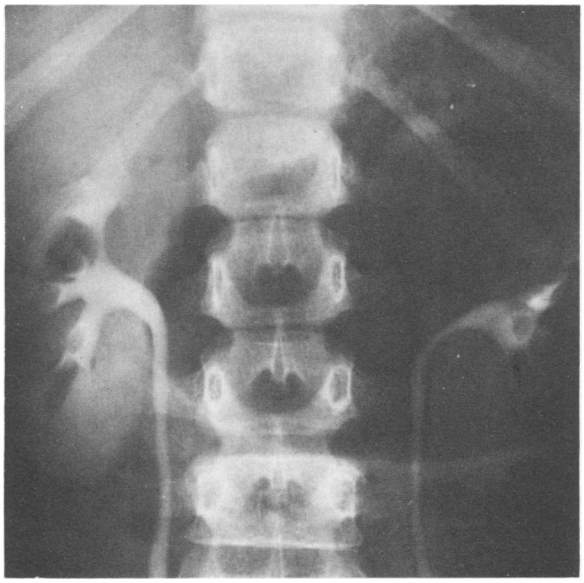

**Figure 2.**—Intravenous urogram film from Case 2: Dilated duplicated collecting system compresses and distorts the lower pole collecting system and gives the appearance of an upper pole intrarenal mass lesion.

upper heminephrectomy and ureterectomy were done and the patient has since been asymptomatic.

## Discussion

Ectopic ureter results from faulty embryogenesis.[1-3] Ectopic drainage can be to any structure derived from the wolffian duct or at sites of ectopic wolffian duct tissue.[4] Ectopic drainage sites in a female patient are the urethra, vagina, cervix, uterus, fallopian tube or rectum.[5] Approximately 80 percent of all ectopic ureters are associated with duplicated collecting systems and the more cephalad system is nearly always the one that drains ectopically.[4,6,7] There is usually high-grade obstruction of the ectopically draining ureter. The vagina is an infrequent site for ectopic drainage (8 percent to 27 percent in several series); the lower portion of the bladder and urethra are more frequently involved.[5,6,8-10] As a general rule, the farther the ectopic ureteral orifice is from its normal location on the bladder floor, the more dilated will be the ureter and the more dysplastic will be its kidney.[11]

The history obtained from a patient with vaginal ectopic ureter is usually characteristic. Urinary incontinence or chronic perineal wetness

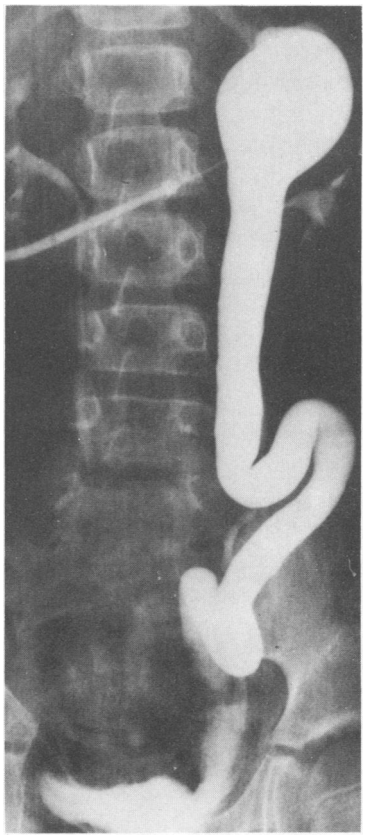

**Figure 3.**—Antegrade pyelogram from Case 2: The dilated duplicated collecting system is seen almost in its entirety. Contrast material in the bladder from previous intravenous urogram obscures the site of entry of the ectopic ureter into the vagina.

after toilet training is highly suggestive of this entity.[3,5,12-14] Such incontinence occurs despite maintaining a normal voiding pattern and may be present only when the patient is upright. This occurs because the involved portion of the kidney functions poorly and the involved ureter is usually dilated and acts as a reservoir in recumbency.[4,9] Other symptoms include vaginal discharge, abdominal pain, back pain and recurrent febrile illnesses.[12-16] Despite the characteristic symptomatology, patients often must consult multiple physicians and have extensive, exhaustive and often duplicated workups.[5,17] Nearly two thirds of patients reach adulthood before their condition is diagnosed and this often results in considerable physical and psychological morbidity.[6,13] Probably most patients with vaginal ureteral ectopia have no positive physical findings, though some will have a palpable abdominal mass or palpable ridge in the vaginal wall caused by the dilated obstructed distal ureter.[9]

Histologically the affected kidney frequently shows severe dysplastic changes. The renal pathology is not necessarily a reflection of primary renal dysplasia (multicystic dysplastic kidney) but can be due to long-standing obstruction or infection or both.[2,11,18]

It is mandatory that an exhaustive evaluation be done to exclude ureteral ectopia whenever a suggestive history exists. Multiple radiologic procedures are available that give an accurate evaluation of this condition. Intravenous urography should be the initial procedure.[17] Duplication with diminished function or nonfunction in the upper pole or unilateral nonfunction or diminished function in the proper clinical setting is very suggestive of this diagnosis and would justify further evaluation. Ultrasonography is useful when a duplicated system with poor function of the ectopically draining system is suspected. This may show the dilated upper pole collecting system and ureter that may not be seen on intravenous urography because of poor function. Also, with unilateral absent or diminished function on intravenous urography, ultrasound evaluation may demonstrate a small kidney and dilated collecting structures. Vaginography[19,20] is a definitive method of evaluation if there is retrograde filling of the ectopic ureter but failure to demonstrate this does not exclude the presence of an ectopic ureter.

These three studies should constitute a minimal evaluation if vaginal ectopic ureter is suggested by clinical history. If ultrasound examination shows a so-called cystic area or hydronephrosis, antegrade injection of contrast medium following percutaneous insertion of a catheter into the renal collecting system can be done. This will usually show the site of ureteral drainage but, if indeterminate, methylene blue or another dye can be instilled and the site of drainage subsequently identified. Cystoscopic and vaginoscopic evaluation are mandatory in attempting to identify the site of ectopic drainage. The ipsilateral absence of a ureteral orifice suggests the presence of an ectopic ureter (see Figure 4).

The surgical management of the ectopic ureter

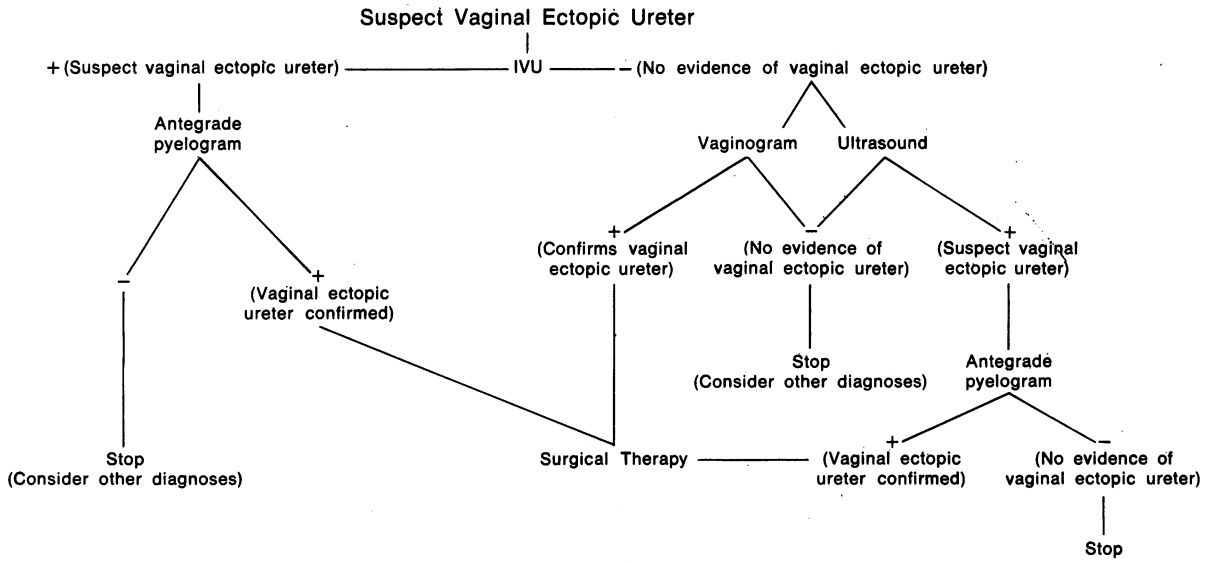


**Figure 4.**—Schematic of diagnostic workup and treatment for suspected vaginal ectopic ureter. + = findings that confirm the diagnosis, – = test fails to confirm diagnosis.

depends on the degree of function in the renal segment drained by the ureter. Most ectopic ureters opening into the vagina drain from renal segments so hydronephrotic and dysplastic that a nephroureterectomy is the best treatment.[3] Most such ureters drain the upper pole of a duplicated system, though occasionally a single ectopic ureter is found draining a dysplastic kidney.[21]

Occasionally an ectopic ureter is encountered with sufficient function remaining to warrant an attempt at salvage. This decision might be made when function in the renal segment is demonstrated on intravenous urography or on renal scan, or by the finding at operation of enough renal parenchyma to suggest future renal functional return. Salvage can be accomplished by a variety of methods involving reimplantation of the ectopic ureter.[5,22]

Some authors suggest removing the upper pole of the kidney and proximal portion of its ureter and leaving the distal ectopic ureter intact because of their fear of devascularizing or otherwise injuring the distal portion of the companion ureter.[3] We have not chosen this approach as it appears to us that when the child becomes a sexually active adult, the retained segment of dilated, poorly draining ureter must certainly serve as a constantly infected perivaginal diverticulum. Other problems may arise from the retained ureteral segment as well.[23] Instead, we remove the ureter to its junction with the vagina and oversew the vagina at that point. Meticulous attention to surgical technique will prevent injury to the adjacent distal ureter.

## Conclusion

Vaginal ectopic ureter results from faulty embryogenesis and results in characteristic symptomatology. Extensive evaluation to exclude the diagnosis is mandatory because failure to diagnose the condition can lead to significant psychologic and physical morbidity. Workup involves a logical sequence of radiographic and endoscopic studies and can be done safely and rapidly. Effective curative surgical therapy is available.

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# *Talcum Powder Pneumoconiosis*

## *Diagnosis by Transbronchial Biopsy Using Energy-Dispersive X-ray Analysis*


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ALTHOUGH COSMETIC TALC is widely used as a dusting powder, it is not generally considered to be a hazard.[1] During its usual use, relatively small quantities are inhaled,[2] and inhaled particles are efficiently cleared by the tracheobronchial tree.

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